CASE REPORT

Cholesteatoma associated with squamous cell carcinoma of the external auditory canal: Case report and literature review

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Abstract

Introduction: Squamous cell carcinoma (SCC) is a rare cancer entity. Only a few cases of squamous cell carcinoma associated with cholesteatoma have been reported in the literature so far. Aim: To describe the case of a squamous cell carcinoma associated with a primary acquired cholesteatoma and discuss the management modalities and the outcome of the disease.

Results: We report the case of a 42-year-old man who presented with a chronic otorrhea and a progressive hearing loss. Otoscopy showed a polypoid external auditory canal. Tonal audiometry revealed a mixed hearing loss with an air conduction threshold of 60 dB. Tomodensitometry and MRI showed erosion of the EAC, lysis of the tympanic bone and the second portion of the facial nerve. Histological examination of the polypoid tissue concluded with a SCC. According to the Pittsburgh staging system, the tumor was classified as T4 N0 M0. The patient underwent a subtotal petrectomy carrying the tumor and sacrificing the facial nerve, a total parotidectomy and a neck dissection of level II–V lymph nodes. Intraoperatively, the tumor tissue was associated with an attic cholesteatoma responsible for ossicular chain destruction. A postoperative radiotherapy was delivered at a dose of 65 Gy. No recurrence or metastases have been reported after a one year follow-up.

Conclusion: SCC of the external auditory canal can mimic cholesteatoma. A precise diagnosis of the disease is important to predict the treatment outcome. Optimal management relies on early surgery and postoperative radiotherapy, thus offering the greatest chance of cure.

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1. Introduction

Squamous cell carcinoma (SCC) of the external auditory canal (EAC) is rare with a prevalence of one per million people.\(^1\)\(^,\)\(^2\) It often mimics other disease entities like chronic otitis media and otitis externa making it a diagnostic challenge. The poor prognosis associated with this carcinoma requires an aggressive surgical management with adjunctive radiotherapy. Cholesteatoma, which is a common diagnosis, is a squamous epithelium trapped in the temporal bone with an extension tendency. Only a few cases of these two entities association have been reported so far.\(^1\)\(^,\)\(^3\)

2. Case report

A 42-year-old man, with no past medical history, presented with a right otorrhea resistant to medical treatment, evolving for 8 months, associated with tinnitus and gradually progressive hearing loss (Fig. 1).

Otoscopy showed a polypoid external auditory canal (EAC), a large marginal perforation of the tympanic membrane with an epidermal invagination. The contralateral ear was normal and nasal endoscopy as well. There was no facial nerve palsy or vestibular syndrome.

Tonal audiometry revealed a mixed hearing loss with an air conduction threshold of 60 dB and an air-bone-gap (ABG) of 40 dB (Fig. 1). CT scan of the petrous temporal bone (Fig. 2) showed erosion of the EAC and opacification of the middle ear space. It also revealed lysis of the tympanic bone and the second portion of the facial nerve.

A biopsy of the polypoid tissue was performed and histological examination concluded to an infiltrating squamous cell carcinoma (SCC) (Fig. 4). Thoracoabdominal computed tomography (CT) performed for initial staging showed no secondary lesion. MRI (Fig. 3) demonstrated a soft tissue mass filling the EAC with an isosignal intensity on T1 and T2 weighted images. It showed an increased enhancement after Gadolinium injection, extending to the mastoid region, the retroauricular soft tissue and the middle ear, without reaching the temporal fossa. The second portion of the facial nerve was involved in the mass.

According to the Pittsburgh staging system, the tumor was classified T4 N0 M0 and the patient was operated on. Intraoperatively, we discovered an attic cholesteatoma with an ossicular chain destruction involving the long process of the uncus and the starboard structure.

Our patient underwent a subtotal petrectomy resecting the tumor, the cholesteatoma and sacrificing the facial nerve whose second portion was invaded by the mass (Fig. 5). Total parotidectomy and neck dissection of level II–V lymph nodes were performed at the same time. The operative cavity was filled with abdominal fat. Surgical margins were negative and definitive histological examination concluded with a SCC of the EAC sparing the parotid gland. Histological assessment of lymph nodes revealed nodal involvement (1N+/16N).

The postoperative course was uneventful and set apart a facial palsy. Our patient received a radiation dose of 65 Gy to the primary tumor site and regional lymph node beds initiated four weeks after surgery. No recurrence or metastases have been reported after a one year follow-up.

3. Discussion

Squamous cell carcinoma (SCC) is a rare cancer entity accounting for less than 0.2% of all head and neck cancers with an incidence of one per million people.\(^3\)\(^,\)\(^10\) It is the most common histological type, representing 80% of temporal bone tumors.\(^5\)\(^,\)\(^10\)\(^,\)\(^13\)\(^,\)\(^17\) Cholesteatoma is one form of chronic otitis characterized by an osteoclastic inflammatory process.\(^3\) Only a few cases of squamous cell carcinoma associated with cholesteatoma have been reported in the literature so far.\(^1\)\(^,\)\(^3\) Owing to the low incidence of the disease, publications and reports in diagnosis and management are rare.\(^10\)

The peak occurs in the seventh decade of life with a masculine predominance.\(^3\)\(^,\)\(^17\)\(^,\)\(^19\) SCC is easily misdiagnosed. As a matter of fact, many diseases such as otitis externa, chronic otitis media and cholesteatoma may lead to confusion with this pathology.\(^10\) In the study of Yin\(^17\), 12.6% of patients with SCC reported a history of chronic otitis media.

Symptoms at initial presentation are usually deafness, otorrhea and ear pain.\(^3\)\(^,\)\(^25\)\(^,\)\(^29\) Bloody otorrhea and otalgia are the

**Figure 1** Tonal audiometry showing mixed hearing loss with a mean ABG of 40 dB.
most common.\textsuperscript{10,17} Facial palsy is seen in 4–20% of cases.\textsuperscript{17,29} Otoscopy may find a fungating mass, a polyp or an invasive tumor.\textsuperscript{29}

Middle ear polyps extending to the EAC are usually associated with cholesteatoma. Nevertheless, tumors such as squamous cell carcinomas should be considered in the differential diagnosis.\textsuperscript{27}

Many authors have implicated cholesteatoma as a causative factor for squamous cell carcinoma.\textsuperscript{8,9,26} Bergmann et al.\textsuperscript{8} detected human papillomavirus (HPV) DNA in 36% of cholesteatoma using polymerase chain reaction. Histologically, lesions were characterized by a bone-destructing form with a papillary growth pattern, as is the case of HPV-associated carcinoma in other anatomical sites as well. Watabe-Rudolph\textsuperscript{9} demonstrated an increased activity of the telomerase in SCC (66%) compared to cholesteatoma (3.4%). This induces a high rate of apoptosis in cholesteatoma and an immortal growth of cancer cells.

Some molecular features could also explain progression from cholesteatoma to SCC. Indeed, Park et al.\textsuperscript{26} examined by immunochemical analysis 40 cases of cholesteatoma and found that survivin was detected in 77% of samples. This protein, also called baculoviral inhibitor of apoptosis, is abundantly expressed in most solid and hematologic malignancies.

Diagnosis imaging modalities are based on CT scan and MRI.\textsuperscript{10,27} They can lead to an early diagnosis and estimate the extension of the disease.\textsuperscript{10}

CT scan is widely used in the preoperative evaluation of chronic otitis media. It shows temporal bone erosion and delineates extension of the lesion.\textsuperscript{27} MRI may provide useful information regarding the tumor staging.\textsuperscript{2,21,27} It is very useful in showing soft tissue lesion.\textsuperscript{12} Gadolinium enhancement on T1 weighted images is seen in benign and malignant tumors, but not in cholesteatomas. Moreover, diffusion weighted images seem very promising since they show high intensity only in cholesteatoma.\textsuperscript{27}

\textbf{Figure 2} Coronal (a and b) and axial (c and d) CT scan of the right temporal bone showing tympanic bone lysis with opacification of the middle ear space.
The Pittsburgh system staging system, based on clinical and radiological findings, was proposed in 1990 and modified in 2000. It is accurate, reliable and reproducible.

When CT scan and MRI are suspicious for malignancy, biopsy is strongly recommended. It may enhance the diagnosis at an early stage and could anticipate insufficient surgery.

Optimal management of SCC remains a matter of debate. For most authors, treatment is based on radical primary surgery and postoperative radiotherapy (RT).

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**Figure 3** Axial T1-weighted (a), Axial Gadolinium enhanced T1-weighted (b), Axial T2-weighted (c), diffusion sequence (d), Coronal Gado T1 MRI (e) and Coronal T2-weighted (f): showing a soft tissue mass filling the EAC with an isosignal intensity on T1 and T2 and an increased enhancement after Gadolinium injection also on diffusion sequences.

**Figure 4** Histological section (HES stain) showing polygonal tumor cells with clear limits, abundant cytoplasm and atypical nuclei. Stroma is fibro-inflammatory with many polynuclear neutrophils. Original magnification ×40 (a) and ×100 (b).
This attitude was adopted for our patient. Early aggressive surgery is advocated by most authors as the standard treatment for this disease. The aim of this surgery is to ensure negative margins and minimize morbidity and mortality. Radical surgery is often difficult and incomplete. This is due to proximity to important organs such as the brain, the internal carotid artery, the cavernous sinus and the parotid gland.

Postoperative radiotherapy plays an important role in local control. Most authors recommend a total dose of 50–70 Gy to control the disease. As metastases are rarely reported, systemic chemotherapy is not routinely used. For Yin, surgery with radiotherapy and/or chemotherapy was the major treatment for tumors stage III and IV while chemoradiotherapy was applied when surgery couldn’t have been performed.

For Hashi, RT with or without surgery was indicated for T1-stage disease while surgery was performed for tumors with bone invasion. Similarly, Ogawa recommends RT for T1 tumors, and surgery followed by radiotherapy for T2 and T3 tumors. Nevertheless, others reserve surgery for T1, T2 tumors and concurrent chemoradiotherapy for T3, T4 tumors.

Prognosis and survival in SCC depends on several factors. Facial nerve palsy, late-stage tumor, lymph node involvement, positive margins, poorly differentiated carcinoma, dural extension and metastases were associated with poorer outcome. Disease free survival rate decreased significantly with stage from 100% for T1 tumors to 21% for T4 tumors.

Early surgery and radiotherapy yields a survival rate of more than 60% after a median follow up of 7 years compared with a survival rate of 33% when salvage surgery or RT alone is performed. Incomplete resection is the most major cause of recurrence and postoperative RT is not efficient in this case. For Ogawa, patients who were treated with preoperative RT tended to have more negative margins than those treated with postoperative RT. In the same study, the use of chemotherapy didn’t influence survival but could enhance the effect of RT. Further studies are required to clarify the role of chemotherapy in these tumors.

Local recurrences and metastases are regarded as therapeutic failures. High recurrence rate (56%) was reported by Taguchi in patients treated with surgery and RT. Regional metastases rate varies between 2.5% and 13% according to some series.

4. Conclusion

Polyps associated with choledempaoma are usually considered as a part of the disease. However, this case report shows that these polyps may be the tip of an iceberg revealing a malignant tumor especially a squamous cell carcinoma. Since the appropriate treatment differs according to the nature of the
mass lesion, preoperative diagnosis is essential. Early referral and aggressive primary surgical treatment with postoperative radiotherapy offer the greatest chance of cure but further studies are required to define the optimal management of these tumors.

Conflict of interest

None declared.

References